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Sclerosing epithelioid fibrosarcoma: case presentation and a systematic review

Ossendorf, C ; Studer, G M ; Bode, B ; Fuchs, B

Abstract: In sclerosing epithelioid fibrosarcoma (SEF), a rare variant of low-grade fibrosarcoma, treatment results and therapeutic options are poorly characterized. We systematically analyzed the data of all 89 patients (43 female, 46 male; mean age, 47 years [range, 14-87 years]) reported in the literature concerning clinical presentation, histopathology, differential diagnosis, treatment, survival rates, and prognosis, and we present an additional case. Information detailing treatment, disease control, and followup was available in 60 (67%), 75 (84%), and 68 patients (76%), respectively. Case history was variable with one-third of patients reporting a painful, enlarging mass. Ten patients (13%) presented with metastases, 23 (31%) had metastases develop after diagnosis, and 28 (37%) had local recurrence. Low cellularity, mild pleomorphism, and sclerotic hyaline matrix of SEF suggest a benign clinical behavior, and cell morphology allows for the wide differential diagnosis of benign, pseudosarcomatous, and malignant proliferations. In addition to surgery, 11 patients (15%) had chemotherapy, 22 (29%) had postoperative radiation therapy, and three (4%) had a combination of both. Twenty-three patients (34%) died from their disease after a mean of 46 months, 24 (35%) were alive with disease, and 20 (31%) were alive without evidence of disease. Patients with SEF of the head and neck had the worst prognosis. **LEVEL OF EVIDENCE:** Level III, prognostic study. See the Guidelines for Authors for a complete description of levels of evidence.

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Sclerosing Epithelioid Fibrosarcoma

Case Presentation and a Systematic Review

Christian Ossendorf MD, Gabriela M. Studer MD,
Beata Bode MD, Bruno Fuchs MD, PhD

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Abstract In sclerosing epithelioid fibrosarcoma (SEF), a rare variant of low-grade fibrosarcoma, treatment results and therapeutic options are poorly characterized. We systematically analyzed the data of all 89 patients (43 female, 46 male; mean age, 47 years [range, 14–87 years]) reported in the literature concerning clinical presentation, histopathology, differential diagnosis, treatment, survival rates, and prognosis, and we present an additional case. Information detailing treatment, disease control, and followup was available in 60 (67%), 75 (84%), and 68 patients (76%), respectively. Case history was variable with one-third of patients reporting a painful, enlarging mass. Ten patients (13%) presented with metastases, 23 (31%) had metastases develop after diagnosis, and 28 (37%) had local recurrence.

Low cellularity, mild pleomorphism, and sclerotic hyaline matrix of SEF suggest a benign clinical behavior, and cell morphology allows for the wide differential diagnosis of benign, pseudosarcomatous, and malignant proliferations. In addition to surgery, 11 patients (15%) had chemotherapy, 22 (29%) had postoperative radiation therapy, and three (4%) had a combination of both. Twenty-three patients (34%) died from their disease after a mean of 46 months, 24 (35%) were alive with disease, and 20 (31%) were alive without evidence of disease. Patients with SEF of the head and neck had the worst prognosis.

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Each author certifies that his or her institution has approved or waived approval for the reporting of this case and that all investigations were conducted in conformity with ethical principles of research.

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C. Ossendorf, B. Fuchs (✉)
Department of Orthopedics, Balgrist University Hospital,
University of Zurich, Forchstr 340, 8008 Zurich, Switzerland
e-mail: bruno.fuchs@balgrist.ch

G. M. Studer
Department of Radiation Oncology, University of Zurich,
Zurich, Switzerland

B. Bode
Institute of Surgical Pathology, University of Zurich, Zurich,
Switzerland

Introduction

Sclerosing epithelioid fibrosarcoma is a rare and poorly recognized but distinct variant of fibrosarcoma (ICD-O code 88 10/3). Since it was originally published in 1995 by Meis-Kindblom et al. in a series of 25 patients [36], 25 reports on 89 patients with a main focus on histopathologic and immunohistochemical features have been reported [1, 2, 4–7, 9, 10, 12–16, 18, 22, 23, 25, 29–32, 36, 40, 42, 43]. These reports suggest SEF is characterized histopathologically by low-grade tumor features: round or oval cells typically arranged in a cord- or nest-like distribution with a collagen background almost effacing the neoplastic cells. Mitotic figures are either scant or absent, and necrosis is uncommon. Sclerosing epithelioid fibrosarcoma is characterized by rearrangement of 10p11 [25, 40] and amplification of 12q13 and 12q15, including the HMGIC gene, a transcriptional activator [16]. Together with low-grade fibromyxoid sarcoma (FMS) and hyalinizing spindle

cell tumor with giant rosettes (HSCTGR) [3], SEF belongs to the family of fibrosing fibrosarcomas [44, 45]. Each of these tumors has distinctive clinical features; they share in common histologic components suggesting a close relationship [3, 20, 21, 42, 43].

The response of SEF to treatment has not yet been fully characterized. Furthermore, the current low-grade classification might be misleading because it disregards the full malignant potential of SEF. The resulting lack of familiarity of treating physicians with SEF might lead to misinterpretation of this rare entity. Thus, the malignant potential of SEF might be underestimated. The potential consequence of this is inadequate therapy leading to an unfavorable treatment result.

Consequently, we systematically reviewed the literature to highlight the following questions: Is there a typical clinical presentation of SEF (patient history, tumor size and location, distant tumor manifestation, and rate of metastasis)? What are the histopathologic features of SEF and what differential diagnoses arise? How were patients with SEF treated and how does this therapy influence overall, disease-free survival and prognosis? Does distant disease depend on tumor size? How do location of the primary tumor, local treatment, and gender influence the prognosis?

Materials and Methods

We performed a systematic review of all SEF studies and case presentations published until December 22, 2007. All publications were derived from NCBI-PubMed (www.pubmed.gov) and Embase (www.embase.com) using the search term sclerosing epithelioid fibrosarcoma. Together, this yielded 34 articles, of which 25 were primarily about SEF. We included all these 25 studies in the current review and analyzed them in detail. The remaining nine studies concern articles about fibromyxoid sarcoma [41] or soft tissue tumor pathology [24, 37], low-grade fibrosarcoma not otherwise specified [28], rhabdomyosarcoma [20], deep

fibromatosis [8], or mention SEF only marginally [21, 27, 33]. Thus, we did not include these studies in our review.

Since 1995, 89 patients (not including our patient) were evaluated in these 25 studies regarding tumor characteristics, treatment, and clinical outcome [1, 2, 4–7, 9, 10, 12–16, 18, 22, 23, 25, 29–32, 36, 40, 42, 43]. Information detailing treatment, disease control, and followup was available in 60 (67%), 75 (84%), and 68 patients (76%), respectively (Supplemental Website Material; a supplemental table is available with the online version of CORR). Including our case, tumors were 8 cm on average and the mean patient age was 47 years (range, 14–87 years) (Table 1). The mean duration between first signs or symptoms and diagnosis was 33 months on average (range, 1 month–13 years). Sixty patients underwent surgery, which included biopsy only ($n = 2$ [3%]); not further specified resection/unspecified surgery ($n = 25$ [42%]); amputation ($n = 5$ [8%]); and wide ($n = 21$ [35%]), marginal, and intralesional excisions ($n = 7$ [12%]). In eight patients, information regarding age was not provided. Sixteen of 82 patients (19%) were younger than 30 years at diagnosis, 30 patients (37%) were 30 to 50 years old, and 36 patients (44%) were older than 50 years. Study quality was judged according to the levels of evidence based on the US Preventive Services Task Force for ranking evidence regarding the effectiveness of treatments or screening as presented by Clinical Orthopaedics and Related Research (<http://www.editorialmanager.com/corr/>).

We tested the influence of gender on prognosis of SEF using the Wilcoxon rank sum test. We used SPSS for Windows (SPSS, Chicago, IL). A value of $p < 0.05$ was considered statistically significant.

Case Report

A 32-year-old woman presented with an 18-month history of increasing swelling at her right distal lateral forearm, which initially was painless. Conventional radiographs

Table 1. Patients' characteristics

Characteristic	Number of patients in whom information was provided per study population	Mean	Quality
Gender	90/90		44 females, 46 males
Age	82/90	47 years (range, 14–87 years)	
History (months)	23/90	33 years (range, 1 month–13 years)	
Size (cm)	78/90	8.3 years (range, 1–25 years)	
Surgery	60/90		Biopsy only ($n = 2$); not further specified resection/unspecified surgery ($n = 25$); amputation ($n = 5$); wide excision ($n = 21$), marginal and intralesional excision ($n = 7$)



Fig. 1 (A) Anteroposterior and (B) lateral radiographs show a punched-out osteolytic lesion surrounded by a sclerotic rim at the mid-diaphysis of the right ulna.

showed a punched-out osteolytic lesion surrounded by a sclerotic rim at the ulna (Fig. 1). Magnetic resonance imaging revealed a 5×1.5 -cm tumor and some edema of the bone marrow (Fig. 2).

We performed a core needle biopsy and after establishing the diagnosis, performed complete gross resection with positive microscopic resection margins. Because of tumor infiltration, the extensor carpi ulnaris muscle and parts of the adjacent muscles were removed. Subsequently, we used a tricortical autologous bone graft from the iliac crest for reconstruction. A 3.5-mm low-contact dynamic compression plate was used to fix the bone graft and to provide stability to the ulna (Fig. 3).

Histologically, there was a sharply demarcated tumor mass in the direct vicinity of the periosteum with infiltration of the cortical bone. The tumor tissue consisted of highly collagenized extracellular matrix with a low density of cells arranged in loose bundles. Most of the cells were elongated with a spindle-shaped nucleus and abundant eosinophilic cytoplasm (Fig. 3A). Focally, particularly at the periphery of the tumor, there were somewhat more cellular areas with plump, epithelioid cells of variable size and individual large, pleomorphic nuclei with prominent nucleoli (Fig. 3B). We detected no mitotic figures or areas of necrosis. Immunohistochemically, no reactivity was detected to any of the tested epithelial, myogenic, or neural markers (actin, desmin, myogenin, S-100, cytokeratin, beta-catenin, CD 31, CD 34, CD 99, CD 117, EMA, bcl-2,

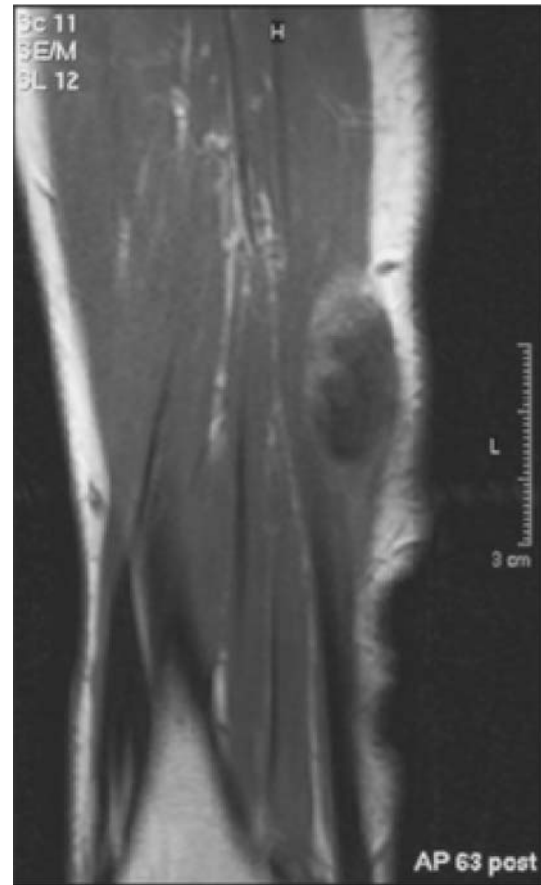


Fig. 2 An MRI scan of the right forearm shows a 5×1.5 -cm tumor extending into the soft tissues and slight bone marrow edema.

MDM-2, MIB-1) with sole positivity for the relatively unspecific, mesenchymal intermediate filament vimentin (Fig. 3C). Thus, we established the diagnosis of a SEF on morphologic criteria.

Subsequently, we administered adjuvant radiation therapy (66 Gy, 1.8 Gy per session). No systemic treatment was given.

Two years postoperatively, imaging studies, including computed tomography of the chest and MRI of the right forearm, confirmed no local tumor and no evidence of systemic disease.

Results

Based on our literature review, the clinical behavior of SEF was generally aggressive with recurrence after resection in 27 of the 90 patients (30%) at the various times of reported followups. One patient had persistent disease and in 15 patients (17%), information regarding recurrence was not provided by the respective authors. Tumor size at diagnosis averaged 8.3 cm (range, 1–25 cm). Most patients had a

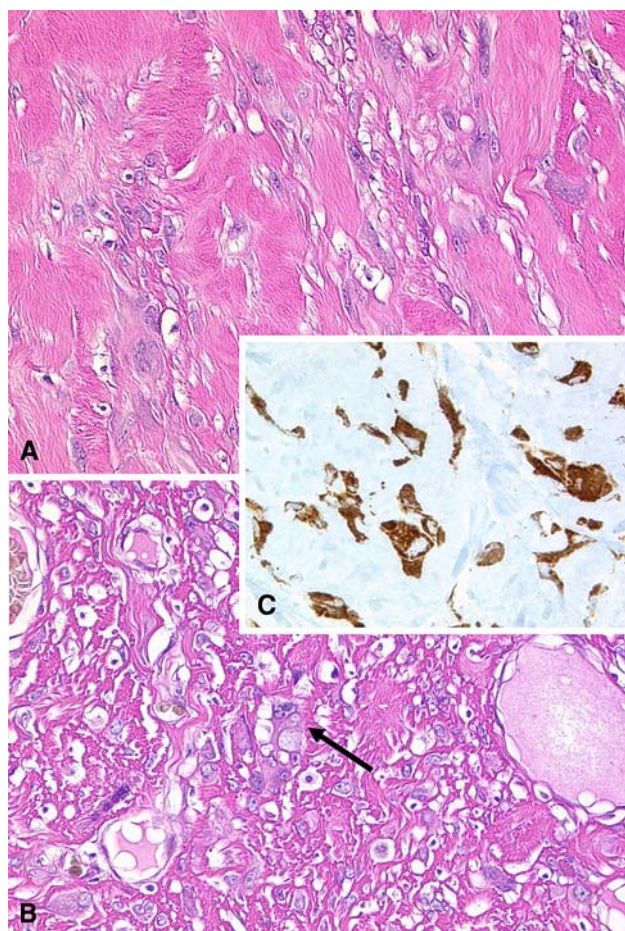


Fig. 3A–C (A) Histologically, the tumor consists of extensive areas with high collagen content and scarce mesenchymal cells (Stain, hematoxylin & eosin; original magnification, $\times 400$). (B) At the tumor periphery, there are areas with higher cellularity and more pronounced cell atypia (arrow) (Stain, hematoxylin & eosin; original magnification, $\times 200$). (C) The tumor cells are immunohistochemically positive for the mesenchymal marker vimentin (Dako, Baar, Switzerland; antibody dilution 1:250) (original magnification, $\times 400$).

variable case history with one-third reporting a painful and enlarging mass, mostly a deep muscular lesion associated with the adjacent fascia or periosteum. The most frequent tumor sites were the lower extremities/limb girdle ($n = 35$ [39%]; average tumor size, 9.37 cm) and trunk ($n = 19$ [21%]; average tumor size, 7.60 cm) followed by the upper extremities ($n = 13$ [14.5%]; average tumor size, 7.65 cm), the head and neck ($n = 13$ [14.5%]; average tumor size, 6.69 cm), and abdominal/inguinal ($n = 10$ [11%]; average tumor size, 9.39 cm). Overall, 75 of 90 patients (83%) were reported to have distant disease at a mean of 36 months after diagnosis. Ten of 37 patients (27%) presented with metastases at the time of the primary diagnosis. The most frequent site of distant tumor manifestation was the lung ($n = 26$ [70%]) followed by osseous lesions to multiple bones ($n = 15$ [41%]) and to the pleura/

chest wall ($n = 4$ [11%]). Two patients had metastases to the scalp (5%). Lymphatic spread, metastases to the breast and to the liver were reported in one patient (3%). Fourteen patients (38%) had metastases to multiple sites. None of the patients with bone involvement had a bone tumor elsewhere.

In SEF, the deceptively low cellularity, mild pleomorphism, and the densely sclerotic hyaline matrix may suggest a benign clinical behavior. Cell morphology allows for the wide differential diagnosis of benign, pseudosarcomatous, and malignant proliferations. The typical morphologic features of sarcomas such as pronounced pleomorphism, infiltrative growth pattern, necrosis, and mitotic activity may be inconspicuous or even absent. These include nodular fasciitis, myositis ossificans, fibromatosis, hyalinizing leiomyoma, fibrous histiocytoma, lobular or signet ring cell carcinoma, sclerosing lymphoma, granulocytic sarcoma, ossifying fibromyxoid tumor, synovial sarcoma, or small cell osteosarcoma.

One patient received preoperative radiation therapy (50 Gy). Twenty-one patients (37%) received postoperative radiation therapy (50.4–70.2 Gy) and 10 of 22 patients (45%) received indicated radiation therapy doses. Ten of 56 patients (18%) received chemotherapy, and three patients underwent combined radiation therapy and chemotherapy. Twenty-seven patients (36%) experienced local recurrence after a mean of 36 months (range, 2–132 months) after diagnosis and treatment. In each age subgroup, there were 50% to 60% of patients who were alive with disease or who had died of disease (DOD).

Distant disease seemed independent of tumor size (nine of 23 tumors [39%] with 5 cm or less, 15 of 33 tumors [45%] with 5–10 cm, and 10 of 22 with 10 cm [45%] or greater metastasized). However, no patient with a primary tumor of 5 cm or less initially presented with metastases. Tumors of patients with metastasis at diagnosis ($n = 10$) were larger (11.3 cm; range, 7.5–17 cm) than tumors that subsequently metastasized ($n = 23$; 7.7 cm; range, 2.5–19.5 cm) and tumors of patients without metastasis ($n = 43$; 8 cm; range, 1–25 cm). Forty-six percent (17 of 37 patients) of patients with metastases experienced local recurrence, and 63% of patients with local recurrence (17 of 27 patients) had distant metastases develop. Seven of 11 patients who underwent systemic chemotherapy had systemic disease develop.

With respect to location, patients with SEF of the head and neck had the worst prognosis with six of 13 patients (46%) DOD followed by SEF of the upper extremities (five of 13 [38%]) and the trunk (five of 19 [26%]). The influence of local treatment could not be assessed adequately because of incompletely reported data regarding surgical procedures, resection status, followup, and missing criteria for radiotherapy.

Sclerosing epithelioid fibrosarcoma of the upper extremities had worse distant control compared with SEF located in the lower extremities with metastasis in six of seven patients. Gender did not seem to influence ($p = 0.069$) local recurrence (13 male and 15 female patients). Followup was available in 16 of 22 patients who received radiation. However, in four and seven of 16 cases, resection status and radiotherapeutic doses were unknown. Twenty-three patients (34%) died from their disease after a mean of 46 months. Twenty-four (35%) were alive with disease and 20 (31%) were alive without evidence of disease.

Discussion

We systematically reviewed the literature about SEF to determine its clinical presentation, histopathology, differential diagnosis, treatment, survival rates, and prognosis. Although histopathologic appearance of SEF suggests low grade, it is a high-grade tumor clinically.

The lack of control groups and prospective design, small sample sizes, and missing treatment details in the reviewed studies limit our interpretations. Because of incomplete data, we could not thoroughly assess the influence of surgery evaluated. The small number of patients in distinct subgroups did not afford adequate power for statistical analysis.

Sclerosing epithelioid fibrosarcoma, together with FMS and HSCTGR, belongs to the family of fibrosing fibrosarcomas. Fibromyxoid sarcoma initially was described by Evans [17] in 1987 and characterized as an indolent, low-grade fibromyxoid neoplasm arising in deep soft tissue with paradoxical aggressive features. Hyalinizing spindle cell tumor with giant rosettes is a similarly rare low-grade fibrosarcoma variant with a very low metastatic potential. In FMS, the local failure rate was 33%, metastases occurred in 42%, and 18% of patients were DOD [18]. Local recurrence rates between 30% and 75% have been observed in several small series with followups ranging from 74% [38] to 100% [7]. In contrast, recurrence of metastasis ranged from 0% [39] to 58% [26]. Died of disease rates ranged from 14% and 33% [16]. Hyalinizing spindle cell tumor with giant rosettes was described for the first time by Lane et al. in 1997 [34], who reported one local recurrence and no metastases in 12 of 19 patients with available followup. So far, six patients with pulmonary metastases have been described [11, 19, 20, 35, 38, 45].

Hyalinizing spindle cell tumors with giant rosettes and FMS have histologic low-grade tumor features and HSCTGR often manifests by an indolent clinical behavior. Based on a series of 56 patients with a mean followup of 3 years, local failure occurred in five (9%) and metastatic

disease in three patients (6%; one HSCTGR, two FMS, but no metastases at diagnosis). One patient was DOD (2%). These data support the hypothesis that FMS and HSCTGR may represent the same entity and should be regarded as low-grade sarcomas [20]. In contrast to HSCTGR and FMS, local recurrence in patients with SEF was seen in 40% and distant disease in 80%, and 67% of patients with local recurrence had distant metastasis develop. Thus, SEF bears malignant potential and therefore appears as the most malignant variant in this family of low-grade fibrosarcomas [20, 21, 34, 42, 43] followed by FMS. This should be taken into consideration when the decision is made on a treatment plan.

Diagnostically, the most challenging problem is to distinguish SEF from undifferentiated carcinoma, which might be impossible based purely on histology. Therefore, immunohistochemical analysis appears mandatory. Specific protein expression or genetic changes have not been described in SEF. The only immunostaining marker consistently reported positive (as in our patient) was vimentin, which is a general marker for soft tissue sarcomas and therefore not specific for SEF [1, 3, 4, 6, 7, 9, 10, 13, 18, 29–32, 36, 40, 43]. Occasional positivity of rare tumor cells for other general markers of soft tissue sarcomas like EMA, S100, or cytokeratin are not useful for diagnosis of a patient with SEF, because these also may be positive in other tumors.

Sclerosing epithelioid fibrosarcoma is a clinically aggressive but histologically low-grade sarcoma with unsatisfactory treatment results. It typically takes 33 months from the first onset of symptoms to diagnosis. Sclerosing epithelioid fibrosarcoma can occur from adolescence to the older years and is localized mainly in soft tissues, but it also can arise as a primary tumor of bone, in the neural system, or in the ovary [40]. Sclerosing epithelioid fibrosarcoma has a predisposition for local recurrence with metastasis primarily to the lung. The role of systemic treatment remains unclear. Consequently, SEF may be treated preferably by resection, including reexcision after intralesional excision. Moreover, preoperative or postoperative radiation as used in other soft tissue sarcomas also should be considered.

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